Pediatric Palliative Care in the 21st Century: Live as Long as Possible as Well as Possible

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Learning Objectives

- Review History of Pediatric Palliative Care (PPC)
- Evaluate “top 10” myths in PPC
- Underscore importance of interdisciplinary team approach

Palliative Care

- **Palliare** (Latin) : to cloak
  - Palliative care is about matching treatment to patient goals.
  - Specialized medical care for children with serious illness
  - Focused on relieving pain, distressing symptoms & stress of a serious illness
  - Appropriate at any age and at any stage, together with curative treatment
  - Goal is to improve quality of life for child/family
Myths, Misconceptions, and Assumptions...

- You are taking care of a seriously ill child. You would not be surprised if she might die within the next few months. You are considering a palliative care consult...

- What are arguments might you hear from colleagues (or family) not to do that?

Assumption # 1:

- The death of a child is a rare event (especially in the USA...)

Death of Children

- 20 children die worldwide every second
- 1,200 children die worldwide every hour
- 29,000 children die worldwide every day
- 15-20 million children would benefit from PPC annually (low-estimate)
- ... and in the US?
How Many Children Would Benefit from PPC

- **UK**: 32/10,000 (high income country)
- **South Africa**: 150/10,000 (upper middle income country)
- **Zimbabwe**: 180/10,000 (low income country)
- No accurate figure globally: ICPCN estimates 15-20 million children worldwide (low estimate)

Mortality 1-4 Year Olds [worldmapper.org](http://worldmapper.org)

PPC ... 2-3 generations ago?
Causes of Death in Children 0-19 years (USA, 2002)

<table>
<thead>
<tr>
<th>Cause</th>
<th>Cases</th>
</tr>
</thead>
<tbody>
<tr>
<td>Birth</td>
<td>18,487</td>
</tr>
<tr>
<td>Life-limiting diseases</td>
<td>15,963</td>
</tr>
<tr>
<td>Unintentional Injury</td>
<td>10,952</td>
</tr>
<tr>
<td>Homicide</td>
<td>2,604</td>
</tr>
<tr>
<td>SIDS</td>
<td>2,050</td>
</tr>
<tr>
<td>Suicide</td>
<td>1,900</td>
</tr>
<tr>
<td><strong>Total</strong></td>
<td><strong>51,956</strong></td>
</tr>
</tbody>
</table>

Life-Limiting Conditions (LLC)

...are those for which there is no reasonable hope of cure and from which children will die before reaching adulthood.

  - White: 27/10,000
  - Chinese: 32/10,000
  - Black: 42/10,000
  - South Asian: 48/10,000

- USA - Age 0-17: 74.3 million children (2014) [http://www.childstats.gov/americaschildren/tables/pop1.asp]: > 237,000 with LLC
  - Prevalence [32/10,000]: 10,800 - 13,700 die/year
  - Mortality [1.5-1.9/10,000]: 15,000 die/year Age (0-24), who would benefit from PPC

Boeing 747-400

416 passengers
US Health Care System...

- USA: > 15,000 children die each year due to life-limiting conditions
- 83 “Boeing 747”
  - one crash every 10 days
  - [every 4.4 days, when including infants]

“Sudden” Death? Advanced Illness Marked by Slow Decline with Periodic Crises and “Sudden” Death

<table>
<thead>
<tr>
<th>Health Status</th>
<th>Time</th>
<th>Death</th>
</tr>
</thead>
<tbody>
<tr>
<td>Multiply relapsed cancer</td>
<td>Crises</td>
<td>Decay</td>
</tr>
<tr>
<td>Cystic Fibrosis</td>
<td></td>
<td>Decay</td>
</tr>
<tr>
<td>Advanced HIV</td>
<td></td>
<td>Decay</td>
</tr>
<tr>
<td>Refractory Seizure Disorder</td>
<td></td>
<td>Decay</td>
</tr>
<tr>
<td>Solid organ transplant recipient</td>
<td></td>
<td>Decay</td>
</tr>
<tr>
<td>Congenital heart disease</td>
<td></td>
<td>Decay</td>
</tr>
</tbody>
</table>

Joanne Wolfe, MD (Boston Children’s Hospital & Dana Farber Cancer Institute), with permission

World Death Rate Holding Steady At 100 Percent

- GENEVA, SWITZERLAND—World Health Organization officials expressed disappointment Monday at the group’s finding that, despite the enormous efforts of doctors, rescue workers and other medical professionals worldwide, the global death rate remains constant at 100

- Death, a metabolic affliction causing total shutdown of all life functions, has long been considered humanity’s number one health concern. Responsible for 100 percent of all recorded fatalities worldwide, the condition has no cure.

Assumption #2:

* Pediatric Palliative Care is usually for children with cancer...

Causes of death in children due to life-limiting conditions

<table>
<thead>
<tr>
<th>Condition</th>
<th>Percentage</th>
</tr>
</thead>
<tbody>
<tr>
<td>Congenital or genetic</td>
<td>16%</td>
</tr>
<tr>
<td>Neurological / neurodegenerative</td>
<td>20%</td>
</tr>
<tr>
<td>Cancer</td>
<td>31%</td>
</tr>
<tr>
<td>Metabolic</td>
<td>9%</td>
</tr>
<tr>
<td>Cardiovascular</td>
<td>12%</td>
</tr>
</tbody>
</table>

Pediatric Cancer: Epidemiology USA

- New cancer diagnosis: > 12,000 children (0-19 years) / year
- Large majority are cured of their malignancy (> 80% of children with cancer are alive 5 years after diagnosis [62% in the mid-1970s]).
- However, children with good fortune to attain cure nonetheless experience considerable suffering
- 1,960 children and adolescents are expected to die due to a malignancy in 2014
Access to PPC?

- Comprehensive palliative care is the expected standard of care for patients with advanced cancer, however access to, and availability of palliative care expertise for the majority of children with life-threatening conditions, is still lacking compared with adult services.

- In the U.S., the vast majority of infants, children, and teenagers with advanced illnesses who are near the end of life do not have access to interdisciplinary pediatric palliative care (PPC) services either in their community or at the nearest children’s hospital.

How well are we doing...?

- Level 1: no known activity (65.6%)
- Level 2: capacity building (18.8%)
- Level 3: localized provision (9.9%)
- Level 4: integration with mainstream providers (5.7%)


Access to PPC?

- 226 hospitals: 162 responded:
  - 112 have PPC program
- Most programs offer only inpatient services, and most only during the work week
Sibling Care

- “What do you gotta do to get some attention in this family...?”


- Siblings interviewed years after the death spoke of wishing they had received more information about their sibling’s death before the terminal period Nolbris, M. and A.L. Hellstrom, Siblings' needs and issues when a brother or sister dies of cancer. J Pediatr Oncol Nurs, 2005. 22(4): p. 227-33.

- During high-school years may be vulnerable to a reconstruction of death-related concepts Noppe, I.C. and L.D. Noppe, Evolving meanings of death during early, middle, and late adolescence. Death Stud, 1997; 21(3) p. 253-75.


Assumption # 3:

* Specifically trained Pediatric Palliative Care specialist are not required...
Breaking Difficult News

Excellent Communication Skills...?

103 Children With Cancer: Last Week of Life

<table>
<thead>
<tr>
<th>Symptom</th>
<th>Percentage</th>
</tr>
</thead>
<tbody>
<tr>
<td>Fatigue</td>
<td>97% (3%)</td>
</tr>
<tr>
<td>Pain</td>
<td>82% (28%)</td>
</tr>
<tr>
<td>Dyspnea</td>
<td>82% (17%)</td>
</tr>
<tr>
<td>Poor appetite</td>
<td>82% (4%)</td>
</tr>
<tr>
<td>Nausea and vomiting</td>
<td>58% (10%)</td>
</tr>
<tr>
<td>Constipation</td>
<td>51% (10%)</td>
</tr>
<tr>
<td>Diarrhea</td>
<td>42% (1%)</td>
</tr>
</tbody>
</table>

<table>
<thead>
<tr>
<th>Symptom treated</th>
<th>Treatment successful</th>
</tr>
</thead>
<tbody>
<tr>
<td>Fatigue</td>
<td>28% (10%)</td>
</tr>
<tr>
<td>Pain</td>
<td>17% (10%)</td>
</tr>
<tr>
<td>Dyspnea</td>
<td>10% (10%)</td>
</tr>
<tr>
<td>Poor appetite</td>
<td>4% (1%)</td>
</tr>
<tr>
<td>Nausea and vomiting</td>
<td>4% (1%)</td>
</tr>
<tr>
<td>Constipation</td>
<td>10% (1%)</td>
</tr>
<tr>
<td>Diarrhea</td>
<td>42% (1%)</td>
</tr>
</tbody>
</table>


Breaking Difficult News

Dr. C. discusses with the parents of a child the poor prognosis of a systemic disease and reviews treatment options ranging from transplant to palliative care.
Subspecialty in Hospice and Palliative Medicine

* American Board of Pediatrics Subspecialty in Hospice and Palliative Medicine

* 2008 first exam given, 46 pediatricians certified

* 2010, 2012, 2014...

* Implications:

  * Requirement for formal training
  * Increase in ACGME/NIH dollars

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Subspecialty in Hospice and Palliative Medicine

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Drugs received by PPC patients

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Symptoms of PPC patients

Assumption # 4:

* Pediatric palliative care starts:
  * when curative treatment stops, and
  * when a child is close to dying
  * and ends at death

*LindyLandzaat*
Palliative care for children and young people with life-limiting conditions is an active and total approach to care, from the point of diagnosis (or recognition) throughout the child’s life, embracing physical, emotional, social and spiritual elements through to death and beyond.

It focuses on enhancement of quality of life for the child/young person and support for the family and includes the management of distressing symptoms, provision of short breaks and care through death and bereavement.

Palliative care no longer means helping children die well, it means helping children and their families to live well and then, when the time is certain, to help them die gently.”

Mattie Stepanek, 1990-2007

Earlier recognition by both physicians and parents that the child had no realistic chance of cure led to a stronger emphasis on treatment to lessen suffering and integrate PPC in pediatric cancer patients.


Religion, Spirituality or Life Philosophy play an important role in the live of most parents whose children receiving PPC


Focus on relieving pain, distressing symptoms & stress of a serious illness

Appropriate at any age and at any stage, together with curative treatment

Goal is to improve quality of life for child/family
Pediatric Palliative Care

- Starts at diagnosis of a life-threatening disease
- Life-threatening AND life-limiting conditions
- Continues through the trajectory of the illness
- Does NOT equal end-of-life care (but includes it)
- Extends beyond the child’s death to the family during bereavement

Hospice vs Palliative Care?

- "Hospice" in USA: Insurance Benefit (Medicare) - unlike rest of the planet...
- All Hospice care is palliative care (!), - but not all palliative care is hospice (!)

Assumption # 5:

* Parents have to choose between “Fighting For a Cure” or “To Give Up”

Continued treatment in face of serious illness

- Parents and pediatric patients may opt for continued treatment of underlying disease even when there is no realistic hope for cure
- Motivated either by hope for a miracle, desire to extend life, or desire to palliate symptoms related to progressive disease
- In discussions of treatment options with families, Wolfe and Grier suggest “The very nature of miracles is that they are rare. However, we have seen miracles, and they have occurred both on and off treatment”

Continued treatment in face of serious illness

- In other words, a child does not have to continue on disease-directed therapy in order to preserve hope, especially when the therapy significantly impacts child’s remaining quality of life.

- Regardless, decisions regarding continued disease-directed therapy need to be carefully considered, weighing the potential for life extension and impact on quality of life.


Pediatric Palliative Care

- Hope for cure and PPC include each other
- Children can graduate from palliative care
- PPC = Advanced management to maintain or improve quality of life
- Despite prevailing assumption: Life-saving care and excellent symptom relief can be provided simultaneously

Assumption # 6:

- Pediatric Palliative Care Means “Giving Up Hope” & “Doing Nothing”
Hope

- Caring for a dying child is emotionally very difficult.
- It may be particularly challenging for physicians and other caregivers to consider the integration of palliative care because this may be perceived as ‘giving up’.
- More importantly, parental loss of a child is certainly considered to be the most difficult type of loss.
- As a result, the emotional cost of recognizing that a child may die impedes planning for optimal care and support.

Hope

- “Tell me, what are you hoping for?”
- Cure?
- Best Quality of Life
- Even when the underlying condition cannot be cured, sophisticated medical technology will be used to control symptoms and improve a child quality of life.
- It is a very active and advanced approach to pain/symptom management and family support.
- “He is not dying because he is not eating...he is not eating, because he is dying...”

An early palliative care intervention (even from the point of diagnosis) = appropriate and beneficial treatments, increased quality of life and may in fact lead to prolonged (!) life.

- RCT, n=151; adult cancer patients receiving palliative care early in their illness lived longer (11.6 months vs. 8.9 months, P=0.02), with better quality of life, including decreased depression.
- Results underscore the need for palliative care early in a serious illness.
- This appear to refute the notion that palliative care means giving up. Patients received palliative care alongside their curative treatment.
- Although this is only one study, it is an exciting one & results are not surprising: PC clinicians regularly see these outcomes in practice - even in pediatric patients.

Assumption #7:

* Pediatric Palliative Care should only occur at a children’s hospital

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Symptom Care Team
Ann Goldman

Number of children with cancer dying at home:

1978-81
before implementation of “Symptom Care Team”:
19 %

1989-90
after implementation of “Symptom Care Team”:
77 %

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Proportion of Children with Cancer Dying at Home

- Washington State, USA: 20%

- Poland: 23%

- Germany: 40%

- England & Wales: 52%
Palliative Home Care


- Children with cancer who received concurrent oncology and palliative home care compared to oncology care alone: Symptom distress experiences were similar. Children enrolled in a PPC home care program appear to have improved quality of life and are more likely to die at home

- Assumption # 8:

  * Increasing the dose of opioids (and/or benzodiazepines) causes respiratory depression and quickens death.
Opioids for Pain & Dyspnea

- "Morphine kills the pain, not the patient" MD killing patient in name of pain relief is not merciful, just incompetent

- An enduring misconception is the belief that in the management of pain and dyspnea, opioids will hasten death and should only be administered as a last resort.
  - This was contradicted in the adult literature...

- ...and our PPC team commonly observes that administering opioids and/or benzodiazepines, together...

Opioids for Dyspnea

- Retrospective cohort study (n=223 adult oncologic patients)

- Mean Survival:
  - < 2-fold increase in their initial opioid dose = 9 days
  - > 2-fold increase in their initial opioid dose = 22 days

Assumption # 9:

- Pediatric Palliative Care takes patients away from primary pediatric specialists
Pediatric Palliative Care Consult

* Complementary
* “How can we help...?”
* May involve second opinion regarding
  * Decision making
  * Symptom management
  * Coordination of care
  * Home care

Assumption # 10:

* A part time (0.2 FTE) physician can certainly address all pertinent PPC issues in a Children’s Hospital

PPC

* PPC applies to all children with life-threatening illness

* PPC Specialists provide:
  * Direct patient consultation
  * Advocacy
  * Education
  * Quality Improvement
  * Research
PPC Example...

* “If you have seen one Pediatric Palliative Care program ... 

* ... you have seen one Pediatric Palliative Care program!

PPC and Cost Saving?


* Ward-Smith P, Korphage RM, Hutto CJ. **Where health care dollars are spent when pediatric palliative care is provided.** Nursing Economics 2008;26:175-8.

Pediatric Palliative Care & Cost Savings?


* Children’s Hospitals and Clinics of Minnesota, Dept. Pain Medicine, **Palliative Care & Integrative Medicine:** 425 children 1-21 years: Home-based PPC or hospice services 2000-2010

* Compare pediatric hospital resource utilization before and after enrollment

* Non-cancer patients: **LOS decrease 38 days, decrease hospital charges $275,000 / patient**
Children’s Hospitals and Clinics of Minnesota (Minneapolis Campus)

- Department of Pain Medicine, Palliative Care & Integrative Medicine

Palliative Home Care (Minneapolis/St. Paul)

- (30 miles) 50 km radius
- Metropolitan area: 3.2 million people
- 24/7; 365 days/year
- 3.5 lead nurses (+ 6 RNs)
- 3 social worker
- 1 child life specialist
- 1 music therapist
- 1 chaplain
- 3 MDs

Pain & Palliative Care & Integrative Medicine Team

<table>
<thead>
<tr>
<th>INPATIENT</th>
<th>CLINICS</th>
<th>COMMUNITY</th>
<th>RESEARCH / QUALITY IMPROVEMENT</th>
<th>EDUCATION</th>
</tr>
</thead>
<tbody>
<tr>
<td>Pain &amp; PPC &amp; IM</td>
<td>Pain &amp; PPC Interdisciplinary</td>
<td>Palliative Home Care &amp; Hospice</td>
<td>Toyota Lean QI</td>
<td>EPEC-Pediatric NIH R25</td>
</tr>
<tr>
<td>Rounding</td>
<td>Pain Interdisciplinary PPC</td>
<td>Perinatal Hospice</td>
<td></td>
<td>Pain Master Class</td>
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<tr>
<td></td>
<td>IM</td>
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<td>IM Master Class</td>
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<td>CAPC PCLC</td>
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<td></td>
<td>MD Fellowship</td>
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<tr>
<td></td>
<td></td>
<td></td>
<td></td>
<td>International Scholar</td>
</tr>
</tbody>
</table>
Pain & Palliative & Integrative Medicine Staff

1.0 FTE Manager
4.2 Physicians
4.0 Nurses
1.0 QI Manager “No Needless Pain”(RN)
5.9 Pediatric Advanced NP / CNS
2 Social Workers
0.6 Child Life Specialist
0.3 Chaplain
2.0 Psychologist
3.0 Administration
2.8 Research
1.8 Massage
0.8 Physical Therapy
3.2 Admin. Assistance

Pain & Palliative Care Team

Continuum

Pain ----- Palliative Care ----- Hospice Care

Home ----- Clinic ----- Hospital

Conclusions:

* Palliative Care is...

* Specialized medical care for children with serious illness
* Focused on relieving pain, distressing symptoms & stress of a serious illness
* Appropriate at any age and at any stage, together with curative treatment
* Goal is to improve quality of life for child/family
* Provided by an interdisciplinary team who work with the patient’s other physicians & health care providers: provides an extra layer of care
* PS.: Morphine & Midazolam do not shorten a child’s life
Conclusions

- Pediatric Palliative Care:
  - Multi--> Inter--> Trans-disciplinary Team

- (1) “How can we help?”
- (2) Then listen…

Further Training

8th Annual Pediatric Pain Master Class | Minneapolis, MN | June 20-26, 2015

Education in Palliative & End-of-life Care [EPEC]: Become an EPEC-Pediatrics Trainer | 2015

Center to Advance Palliative Care (CAPC) - Pediatric Palliative Care Leadership Center (PCLC)

Training | 2015

Blog: http://noneedlesspain.org

Caring for Children at the End of Life
Group 1

Life-threatening conditions for which curative treatment may be feasible but can fail, where access to palliative care services may be necessary alongside attempts at curative treatment and/or if treatment fails.

Examples: cancer, irreversible organ failures of heart, liver, kidney

Royal College of Paediatrics and Child Health (UK) & Association for Children with Life-threatening Conditions and their families (ACT)

Group 2

Conditions where premature death is inevitable, where there may be long periods of intensive treatment aimed at prolonging life and allowing participation in normal activities

Examples: cystic fibrosis

Royal College of Paediatrics and Child Health (UK) & Association for Children with Life-threatening Conditions and their families (ACT)

Group 3

Progressive conditions without curative treatment options, where treatment is exclusively palliative and may commonly extend over many years.

Examples: Batten disease (neuronal ceroid lipofuscinosis), mucopolysaccaridoses, muscular dystrophy

Royal College of Paediatrics and Child Health (UK) & Association for Children with Life-threatening Conditions and their families (ACT)
Irreversible but non-progressive conditions with complex healthcare needs leading to complications and likelihood of premature death.

Examples: severe cerebral palsy, multiple disabilities following brain or spinal cord insult

Royal College of Paediatrics and Child Health (UK) & Association for Children with Life-threatening Conditions and their families (ACT)

How is PPC different from adult palliative care?

Compared with adult palliative care, PPC:

- Serves smaller numbers of patients and families
- Serves a broad spectrum of illnesses, including many rare diseases, often requiring involvement of a number of medical disciplines
- Serves children with unpredictable illness trajectories
- Is provided for the duration of the child’s illness
  - Overlap with chronic or curative care
  - PPC becoming more prominent as the child transitions from chronic to terminal illness
  - PPC may last anywhere from days to more than a decade
- May include life-extending interventions such as noninvasive respiratory ventilation, blood transfusions, or parenteral nutrition, which may also help meet important palliative needs

How is PPC different from adult palliative care?

Compared with adult palliative care, PPC:

- Often involves a multitude of decision makers, including patients, parents/caregivers and clinicians
  - Results in the need for highly nuanced communication strategies.
- Always involves the added emotional burden of a child possibly dying before the parent
  - For parent this is unnatural and “out of order”
  - Clinician may experience this as a failure
- Involves challenges associated with patients’ ages, which range from infants to young adults
  - Requires considerable flexibility in styles of communication, symptom assessment, and treatment modalities
- Requires knowledge of the increased complexity of pharmacokinetics and pharmacodynamics in children and adolescents both with regard to prescribing and monitoring pharmacotherapy
Palliative care is an approach that improves the quality of life of patients and their families facing the problem associated with life-threatening illness, through the prevention and relief of suffering by means of early identification and impeccable assessment and treatment of pain and other problems, physical, psychosocial and spiritual.

Palliative care:

- Provides relief from pain and other distressing symptoms;
- Affirms life and regards dying as a normal process;
- Intends neither to hasten or postpone death;
- Integrates the psychological and spiritual aspects of patient care;

http://www.who.int/cancer/palliative/definition/en

Palliative care:

- Offers a support system to help patients live as actively as possible until death;
- Offers a support system to help the family cope during the patient's illness and in their own bereavement;
- Uses a team approach to address the needs of patients and their families, including bereavement counseling, if indicated;
- Will enhance quality of life, and may also positively influence the course of illness;
- Is applicable early in the course of illness, in conjunction with other therapies that are intended to prolong life, such as chemotherapy or radiation therapy, and includes those investigations needed to better understand and manage distressing clinical complications.

http://www.who.int/cancer/palliative/definition/en

Palliative care for children:

- Active total care of the child's body, mind and spirit, and also involves giving support to the family
- Begins when illness is diagnosed, and continues regardless of whether or not a child receives treatment directed at the disease
- Demands that health providers evaluate and alleviate a child's physical, psychological, and social distress
- Requires broad interdisciplinary approach
- Includes the family and makes use of available community community resources; it can be successfully implemented even if resources are limited
- Can be provided in tertiary care facilities, in community health and hospice centers, and in children's homes
- Should be developmentally appropriate and in accordance with family values

http://www.who.int/cancer/palliative/definition/en